Paradoxical embolism in Ebstein’s anomaly

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A 30-year-old woman presented with sudden onset left flank pain 10 days following an uncomplicated delivery. She was normotensive with an electrocardiogram showing sinus-rhythm and pre-excitation (Panel A). Abdominal-computed tomography revealed a large wedge-shaped infarct in the left kidney (Panel B, arrows). Lower-extremity doppler-imaging showed small bilateral deep-vein thromboses and anti-coagulation was initiated. Echocardiography imaging showed small bilateral deep-vein thromboses and anti-coagulation was initiated. Echocardiography suggested apical displacement of the tricuspid septal-leaflet consistent with Ebstein’s anomaly. Cardiac magnetic resonance imaging demonstrated a small well-functioning right ventricle, 5 cm apical displacement of the tricuspid septal leaflet with mild tricuspid regurgitation (Panel C, arrow, Supplementary material online, Video S1), and a small secundum atrial-septal defect (ASD) with left-to-right shunting (Qp/Qs = 1.3) (Panel D, arrow, Supplementary material online, Video S2). These findings were highly suggestive of paradoxical embolization through the ASD and she was referred for percutaneous closure. Intraprocedural transesophageal 2D and 3D images of the atrial septum were obtained before (Panels E and F, arrows, Supplementary material online, Videos S3 and S4) and after successful closure with a 17mm-Amplatzer device. (Panels G and H, arrows, Supplementary material online, Videos S5 and S6).

The clinical presentation of Ebstein’s anomaly is highly variable—depending on anatomic severity, hemodynamics, and degree of interatrial shunting. The majority of patients have shunting through a secundum ASD or patent-foramen ovale. Paradoxical embolism maybe an indication for ASD closure. However, there are concerns that isolated ASD closure may increase tricuspid regurgitation and cause deterioration of right ventricular function. Limited data suggest that percutaneous closure can be performed safely in selected patients with mild-to-moderate tricuspid regurgitation and left-to-right shunting. In patients with right-to-left shunting and cyanosis, haemodynamic monitoring during test occlusion of the defect—at rest and under catecholamine stimulation—may help evaluate the feasibility of closure.

Supplementary material is available at European Heart Journal online.

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